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Case Report

Unusual MRI characteristics of Posterior Cranial Fossa Dermoid Cyst Associated with Klippel-Feil syndrome: A Case Report and Literature Review.

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Abstract
Intracranial dermoid cysts are rare inclusion cysts characterized as well-defined cystic lesions with typically distinct Magnetic Resonance Imaging (MRI) features. However, in rare cases, their imaging features can be atypical because of their variable components. This case report highlights unusual radiological findings of a posterior cranial fossa dermoid cyst that deviates from typical presentation patterns and reveals a rare association between these lesions and Klippel-Feil syndrome (KFS). We detail the case of a 50-year-old female patient who presented with progressive weakness, persistent headaches, and deteriorating vision. MRI of the brain displayed a 6X4.5 cm well-defined cystic lesion in the posterior fossa, showing hyperintensity on T1-weighted images (T1WI) and profound hypointensity on T2-weighted images (T2WI), along with cervical vertebrae fusion indicative of KFS. These atypical imaging findings of posterior cranial fossa dermoid cyst challenge conventional diagnostic patterns and underline the necessity for further exploration of these lesions to avoid misdiagnosis. Also presence of posterior cranial fossa dermoid cysts should be considered in a patient diagnosed with KFS.
Key words: Dermoid cyst**,** Magnetic Resonance Imaging, Klippel-Feil Syndrome.

Introduction

Dermoid cysts are benign congenital lesions that develop during embryogenesis between the third and fifth weeks of gestation. These slow-growing lesions arise from the entrapment of ectodermal cells during the closure of the neural tube, resulting in encapsulated lesions within the central nervous system [1]. They are characterized by the presence of ectodermal elements, including hair follicles, sweat glands, and sebaceous glands [2]. Although they can manifest either intra- or extracranially, intracranial dermoid cysts are particularly rare, representing less than 0.5% of all intracranial tumors. These lesions are frequently present in individuals within the first four decades of life, with a slight predominance observed in females [1]. Clinically, although intracranial dermoid cysts are often discovered incidentally through imaging studies, they can also present with symptoms such as headaches, seizures, and focal neurological deficits, depending on their size, location, and risk of rupture. The imaging characteristics of these lesions are crucial for accurate diagnosis; intracranial dermoid cysts typically exhibit a hypodense appearance on CT scans and hyperintensity on T1-weighted MRI (T1WI), with heterogeneous signal intensity on T2-weighted images (T2WI). Techniques such as fat-suppressed MRI sequences are endorsed for accurately diagnosing ruptured cysts. Additionally, enhancement is an uncommon criterion that may help distinguish these cysts from other intracranial tumours[2]. And non-complicated cases are never associated with brain edema [3]. Posterior fossa dermoid cysts and Klippel-Feil syndrome (KFS) represent a rare but clinically significant association. This unique relationship highlights an intriguing association of embryological development and congenital anomalies. KFS is characterized by the congenital fusion of cervical vertebrae and might be associated with other congenital anomalies [4]. The association of intracranial dermoid cysts in individuals with KFS suggests a distinct pathophysiological link that deserves further investigation. Thus, this case highlights unusual MRI findings of an intracranial dermoid cyst that depart from typical presenting patterns and reveals a rare association between these lesions and KFS.

Case report:

A 50-year-old woman presented with concerning symptoms, including persistent headaches, vomiting, imbalance, and frequent falls. Notably, she had not experienced any recent head trauma or fever and did not have a history of any previous surgical interventions or family history of similar conditions. At the time of her evaluation, all laboratory tests returned normal results. MRI of the brain revealed a 6X4.5 cm well-defined extra-axial midline cystic lesion within the posterior fossa with a small midline occipital transdiploic linear channel at the closely adjacent occipital bone. The lesion was hyperintense on TW1 and extremely hypointense on T2WI; no diffusion restriction, contrast enhancement, or perilesional edema was observed, and no signs of cyst rupture (Figure 1). On T2* susceptibility images, there is low signal within the wall and the mural nodule, which may signify an area of calcifications. The cyst compressed the fourth ventricle, causing mild supratentorial obstructive hydrocephalus. Furthermore,

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sagittal MRI of the cervical spine revealed fusion of cervical vertebrae, consistent with KFS Figure 2.

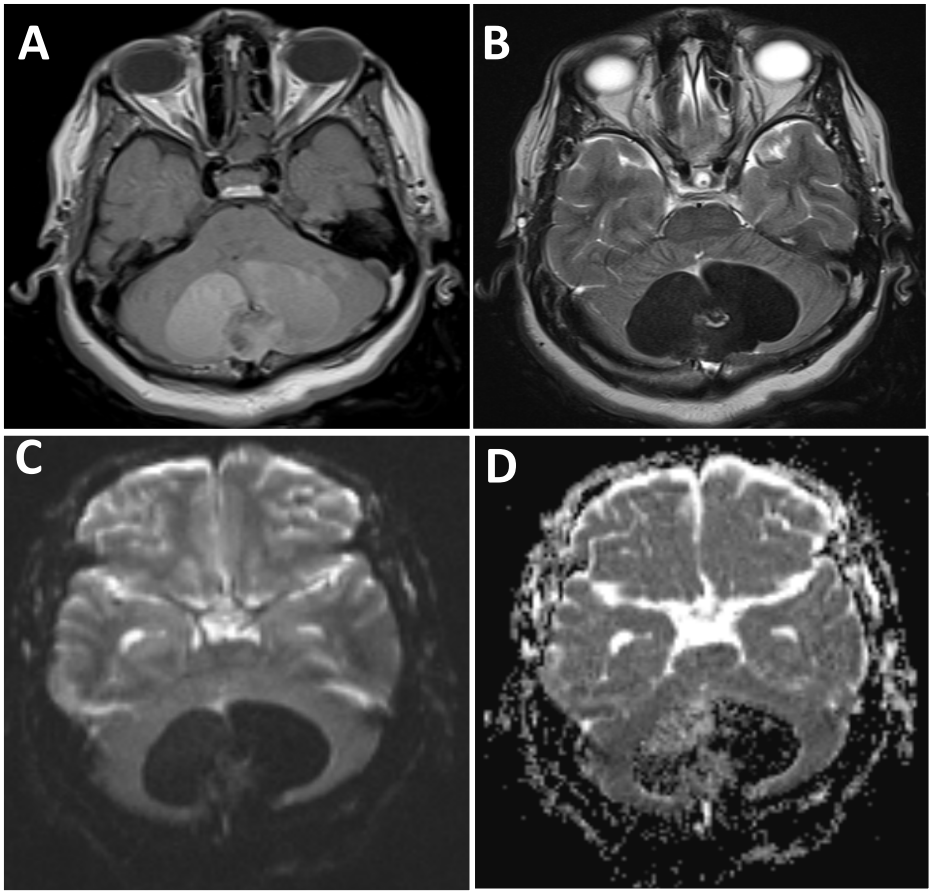

Figure 1.: axial brain MRI showing a well-circumscribed lesion at the midline posterior fossa, which is profusely hypointense on T2-weighted sequences and hyperintense on T1-weighted sequences with a small midline transdiploic channel (A, B). DWI and ADC show no diffusion restriction of the lesion (C, D).

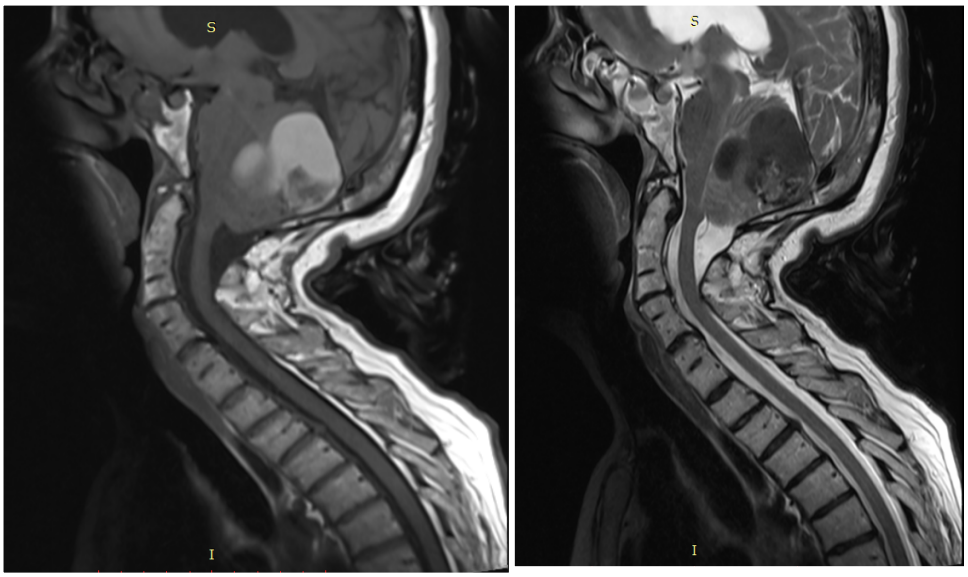

Figure 2 sagittal T1 and T2 MRI cervical spine shows fusion of cervical vertebrae consistent with Klippel-Feil syndrome.

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Discussion

This report presents a rare case of a 50-year-old female patient with a posterior cranial fossa dermoid cyst characterized by unusual MRI signal characteristics, specifically hyperintensity on T1WI and marked hypointensity on T2WI, concomitant with cervical vertebrae fusion consistent with KFS. Dermoid cysts are usually well-defined, often with wall calcification [1]. Their high lipid content gives them distinctive imaging characteristics, appearing hypodense on CT and hyperintense on T1-weighted MRI [2]. On T2-weighted MRI, they may appear heterogeneous or hyperintense[5]. However, in the current case, the intracranial dermoid cyst presented with hyperintensity on T1WI and was profoundly hypointense on T2WI. To our knowledge, such imaging findings are extremely rare, with only a few cases reported worldwide (Table 1) [6]. This underscores the need for further investigation into the variability of the MRI signal in these lesions.

Abbas et al. discuss in their review previously reported cases of posterior fossa dermoid cyst with atypical imaging features, with only six cases where intracranial dermoid cysts exhibited atypical hyperintense T1 and hypointense T2 signal on MRI, which may be mistaken for other lesions. This finding is significant because it highlights the potential for diagnostic errors due to the rarity and unexpected nature of such imaging characteristics [6].

Table 1: Summary of previously reported cases of posterior cranial fossa dermoid cysts exhibiting characteristic hypointense T2 Signals.

Study	Age	Sex	MRI T1	MRI T2	DWI	Mural nodule
Brown *et al.*, (2001)	18	F	Hyperintense	Hypointense	-	positive
Neugroschl *et al.*, (2002)	73	M	Hyperintense	Hypointense	-	-
Li *et al.*, (2011)	14	M	Hyperintense	Hypointense	-	positive
Sathiaprabhu *et al.*, (2020)	37	M	Hyperintense	Hypointense	Low	positive
Abbas *et al.*, (2022)	35	F	Hyperintense	Hypointense	-	positive
Delgado-Muñoz *et al.*, (2022)	39	F	Hyperintense	Hypointense	Low	Positive
Our case	50	F	Hyperintense	Hypointense	Low	Positive

While the precise etiology behind the observed atypical MRI signals, specifically hypointensity on T2WI, remains uncertain, several factors may contribute to these unexpected imaging characteristics. These could include lipid saponification, the presence of keratinized debris, suspended microcalcifications, liquefied cholesterol, increased protein content, and/or hemosiderin resulting from previous internal hemorrhage [2]. The link between dermoid cysts of the posterior cranial fossa and cervical fusion anomalies, specifically KFS, was first reported by Love and Kernohan in 1936, and since then, only 25 additional cases have been documented in medical literature by Zhang et al [7]. The association of these two entities is believed to be related to disturbances in mesodermal development during early embryogenesis, which affects both vertebral segmentation and posterior fossa formation[8]. KFS is marked by the congenital fusion of cervical vertebrae attributable to improper segmentation of the paraxial mesoderm and may plausibly affect ectodermal components during neural tube development [4]. Simultaneously, dermoid cysts are thought to develop from the inclusion of ectodermal components, such as skin and sebaceous glands, into the neuroectoderm during neural tube closure, particularly in instances of abnormal morphogenesis like KFS [7]. Therefore, the association between KFS and posterior fossa dermoid cysts suggests a shared embryological disruption in spinal and cranial development, connecting the two developmental disorders. This highlights the need for meticulous evaluations of KFS patients for potential intracranial dermoid cysts due to their rare yet significant clinical implications.

Conclusion

This case report highlights the diagnostic nuances of a dermoid cyst in the posterior cranial fossa, which presented with unconventional MRI signal characteristics and a misleading radiologic appearance. The case underlines the importance of considering dermoid cysts in differential diagnoses even when classical imaging features are absent, and may help radiologists avoid misinterpretation in similar scenarios. It also underlines a rare but important association between these lesions and KFS, which calls for further investigation into their shared embryological origin.

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The article has not been previously presented or published, and is not part of a thesis project.

Conflict of Interest

There are no financial, personal, or professional conflicts of interest to declare.

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